INFILTRATING ANGIOLIPOMA: A REVIEW OF THE LITERATURE AND A CASE REPORT WITH A FIVE YEAR FOLLOW-UP

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Infiltrating Angiolipoma: A Review of the Literature And A Case Report with a Five Year Follow-Up

by

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Dartmouth College B.A. 1971

A thesis submitted to the Department of Orthopaedic Surgery, in partial fulfillment of the requirements for the degree of Doctor of Medicine.

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April, 1976

New Haven, Connecticut

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Introduction

The objective of this thesis is to describe the clinical entity, infiltrating angiolipoma through an analytical review of the literature. Representative studies will be compared with a discussion of the clinical presentation, diagnosis, treatment and prognosis. To this end, a new case of infiltrating angiolipoma will be presented with emphasis on the similar and divergent aspects relative to cases previously reported.

Infiltrating angiolipomata is defined as unencapsulated lipomatous lesions containing a rich vascular component involving skeletal muscle as well as deep and peripheral soft tissue. In addition, they tend to recur after incomplete local excision.[10] The tumor is sometimes called a vascular lipoma depending on its composite ratio of fat cells, capillaries, and stroma. The nodule of angiolipoma may be interpreted grossly as lipoma but the degree of vascularity is much greater than that of simple lipoma when examined microscopically.[48] Clinically, the characteristics distinguishing it from common lipomata are its painful lesions.[10] This appears to be a variable finding and will be further discussed.

A lipoma is a benign tumor composed of fatty tissue that forms firm, encapsulated, elastic lesions. This consistency may over time change because of fibrosis, necrosis, or cal-

cification. The lesion is readily excised because it generally arises in the sub-cutaneous tissue. However, occurrences of the lesion in deeper structures and organs have been documented by different authors.[1,4,7,12,15,27,29,32,34,36,40,53] Ewing introduced in 1940 the entity of encapsulated angiolipoma which he termed lipoma telangiectaticum.[14] He observed that the lipomata have a striking connection with nutrient blood vessels, with each of the multiple lobules growing about a branch of the main vessel. The extensive overgrowth of these nutrient vessels, Ewing concluded, was responsible for the formation of vascular fatty tissues such that it was sometimes difficult to distinguish it from the hemangioma.

Reports of infiltrating angiolipomata per se are relatively few. Since Paget's description of an infiltrating lipoma in 1853,[33] 191 cases have been reported. Most of these cases appear prior to 1929. In the more recent literature, since 1929, only 27 cases have been reported. The sparsity of reported cases could be a consequence of the actual rarity of the tumor, or its simply not having been diagnosed as such. These two factors alone point out the necessity for more thorough and detailed case reports and reviews. It is most likely a combination of the two factors mentioned as well as others which contribute to the small number of cases reported.

Review of the Literature

There are no reports of infiltrating angiolipoma in the literature prior to 1966. However, the entity of infiltrating lipoma constitutes all the cases of unencapsulated lipomatous lesions reported between 1853 and 1966. The first case of a lipoma infiltrating muscle was apparently reported by James Paget in 1853. He described the lipomatous growth as having "distinct prolongations that passed between the fasciculi of the trapezius muscle."[33] Three additional cases of infiltrating lipoma were described by Broca in 1869.[10]

Pitres, in 1873, was the first to describe the presence of skeletal muscle within the tumor substance. In 1903, Dertinger collected 127 cases from the literature and included 12 personal cases in a study on deep-seated lipoma of the extremities. He concluded that the differential diagnosis was difficult.[13]

In 1929, Behrend reviewed the literature of 190 cases which had been collected to date and reported 2 personal cases.[3] Dionne, however, points out that the collection was not complete since Morestin's case of 1897 was omitted.[13] In Behrend's study, the objective was to dispel the confusion that existed regarding intra-intermuscular lipomata. In his classification, he defined intermuscular lipoma as being encapsulated lesions

that are found between muscle groups. Intramuscular lipoma or the other hand was defined as diffuse lesions infiltrating muscle substance.

Adair reported in 1932 an infiltrating lipoma in a boy's arm. However, Dionne contends that Hoffman's case report in 1941 was a more convincing example of infiltrating lipoma.[13] This lesion was found in the calf of a 42 year old woman. Regan, likewise, in 1946 reported a case of infiltrating lipoma involving the calf muscle. In 1947, Oosthuisen and Barnetson presented two middle aged men with progressive enlargement of a mass lesion in the right foot since birth. Extensive bony invasion and joint destruction were noted on roentgenogram and were thus confirmed on pathologic study of the amputated legs. The calf muscle of amputated specimins were also found to be diffusely infiltrated by the lesion.[31]

Greenberg et al., in their study in 1963, described two cases of infiltrating lipoma of the thigh which are summarized in Table 1.[18] In their study, they included a discussion on differences between inter-intramuscular infiltrating lipomata. The two cases were meant to serve as illustrations of both types of muscle involvement.

TABLE I Greenberg et al.: Case Summaries of Infiltrating Lipoma

	Case 1	Case 2
j) location	thigh	thigh

	Case 1	Case 2
ii) tenderness/pain	yes	no
iii) bony involvement	no	no
iv) treatment	sharp dissection	excision
v) recurrence	--	--
vi) age of onset	50	67
vii) sex	F	F
viii) follow up period	1 year	1 year

They concluded that infiltrating lipoma occurred within the deep muscles of the thigh in patients within the fourth to sixth decade. They also noted that the intermuscular lesions were more common and readily excised than the intramuscular lesions. They recommended meticulous histological examination to exclude liposarcoma. Finally, they advised complete local excision to prevent recurrence of tumor.

It was not until the study of Enneking et al. in 1966 that infiltrating angiolipoma was described as a distinct clinicopathologic entity.[10] These investigators reported six personal cases, the characteristic features of which are summarized in Table II. These investigators looked at the histologic criteria which they believed essential for the diagnosis of infiltrating angiolipoma. They emphasized the process of infiltration and displacement of muscle fibers by mature fat cells. Furthermore, they noted no signs of mitosis or lipoblastic activity among the fat cells. Another histologic feature stressed by the authors is that this fatty tissue be combined with variable collagenous and angiomatous components.

According to these workers, the angiomatous pattern seen in infiltrating angiolipoma is distinct from that seen in the ordinary lipoma. The vessels found in infiltrating angiolipoma are extremely variable in caliber, and are neoplastic in appearance. That is, the veins were a collection of thin walled cavernous vessels with anastomatic branching. The arterioles were recognizable by their extremely thickened walls secondary to smooth muscle hyperplasia.

In spite of the locally malignant bahavior of angiolipoma the investigators were convinced that the lesions were benign congenital malformations. These lesions, they believed, were derived from sequestrated multivalent mesenchymal elements. This etiologic postulation stems from the fact that "adipose cells in the fetus are derived from specialized vascular tissue, while in the adult, their source is from mesenchymal cells."

TABLE II Enneking et al.: Case Summaries of Infiltrating Angiolipoma

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
) location	left thigh	left knee	right calf	right knee	left heel	paraspinal L_{1-4}
i) pain	no	yes	yes	no	yes	yes
ii) bony involvement	no	no	yes	no	no	yes
v) treatment	rad. exc.	rad. exc.	partial exc. x2	rad. exc. x2	rad. exc. x2 then radiotherapy	rad. exc.
) recurrence	no	no	yes	yes	yes	no

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
vi) age of onset	27	$6\frac{1}{2}$	27	$3\frac{1}{2}$	26	
vii) sex	M	F	F	M	F	F
viii) follow up period	4 years	3 years	5 years	8 months	13 months	37 months

Abbreviations used

1. rad. exc. = radical excision
2. partial exc. = partial excision
3. x2 = twice

Stimpson (1971) reported seven new cases of infiltrating angiolipomata (Table III).[47] This investigator also observed that gross inspection alone was not adequate to make the diagnosis of angiolipoma. The vascularity itself may not be visually conspicuous and signs of hemorrhage may be absent. Of the seven cases that he reported only one, which will be described more fully, had grossly apparent vascularity and hemorrhage at time of excision. Thus according to this author, the diagnosis must be validated on microscopic examination. He remarked that the histology showed irregular clumps of fat envelopes separating muscle fibers and resulting atrophy and death of muscle. He also observed that the percentage of angiomatous component was variable in infiltrating angiolipoma and that tortuosity was a distinguishing property of the vessels.

TABLE III Stimpson: Case Summaries of Infiltrating Angiolipoma

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7*
i) location	Pectoralis major	Thigh	Scapula	Neck strap muscle	Groin	Deltoid	Left side
ii) pain	no	yes	no	yes	no	no	no
iii) bony involvement	no	no	no	no	no	no	no
iv) treatment	partial then rad. exc.	rad. exc.	rad. exc.	exc. $\bar{p}$ radiation	exc. $\bar{p}$ radiation	sub-total then rad. exc. & radiotherapy	exc.
v) recurrence	yes	no	no	no	no	yes	yes
vi) age	2 years	51	53	56	56	63	23
vii) sex	F	M	M	F	M	F	M
viii) follow up period	9 months	18 months	18 months	12 months	3 years	18 months	3 months

* A fuller description of this case is seen following.

Abbreviations used

1. $\bar{p}$ = followed by
2. rad. exc. = radical excision

Case vii A 23 year old male presented with two tender masses on his left side of two years duration. They were then excised by diathermy and sharp dissection. After 21 months, he had a recurrence of two tumor mass: one in the operative scar and another in the left iliac fossa. He was readmitted for surgery. A lipomatous mass with apparent tortuous vascularity was excised from the subcutaneous tissue. A second mass was

also seen within the external oblique muscle. This mass had a vascular channel running into the internal oblique. However, excision of this vascular channel was not complete. Within three months of the second surgery, the patient again presented with a tender soft-tissue swelling on the midaxillary line. He was offered radiotherapy but he refused it.

Stimpson compared the clinical features of angiolipoma with that of the corresponding lipoma, found the former to be generally more tender and painful. Furthermore, he noted that clinical diagnoses of angiolipoma could be suspected if the edge of the mass remained ill-defined when the associated muscle was contracted.

Stimpson compared his cases with those of the 1966 study by Enneking et al. and brought into question two ideas that were presented in that study. The first concerns location of involvement. While 80% of Enneking's cases involved the lower limb, Stimpson was able to demonstrate a more varied distribution. (Table III). The second concerns age of onset. The author pointed out that Enneking's cases were loaded towards the first three decades of life whereas five of his personal cases were in the sixth decade.

Stimpson shared the prevailing opinion that wide local surgery was necessary to counter the recurrence of the lesion. However, he stated that the limitation with radical surgery stemmed from its being mutilating as well as being dangerous

to life where there was invasion of contiguous vital structures. These workers therefore advised surgery be supplemented with radiotherapy if either of these two situations ever occurred.

The most current work in the literature comes from Dionne and Seemayer.[13] Their study is a comprehensive review of infiltrating lipomatous lesions, including angiolipomata. These workers, however, included in their analysis a case reported by Davis which does not qualify as a true case of infiltrating angiolipoma. This diagnosis is not consistent with the description given by Davis himself;[11] that is, there is no histological description of any vascular element in the case reported.

The study is also an analysis of cases collected since Adair's case report in 1932. To their study, they added seven new cases of infiltrating lipomata (Table IV). The criteria used for histologic diagnosis of angiolipoma in this study did not differ from that of Enneking or Stimpson. In spite of the local aggressiveness of these lesions, the authors stressed that malignant transformation into liposarcoma has never been reported in either type of infiltrating lipomatous growth. Nevertheless, they recommend that meticulous histologic examination be done to eliminate coexisting liposarcoma. The prevailing opinion concerning the origin of liposarcoma is that it arises de-novo. Therefore, the authors concluded that lipo-

sarcoma coexisting with angiolipoma should be considered an incidental finding and never a product of malignant transformation. To this end, their second case is an example of an instance in which repeated histologic study revealed a coexisting liposarcoma (Table IV).

TABLE IV Dionne and Seemayer: Case Summaries of Infiltrating Lipoma

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7
i) location	leg	leg	triceps	thigh	forearm	leg	thigh
ii) tenderness	no	no	no	no	no	no	no
iii) bony involvement	no	no	no	no	no	no	no
iv) treatment	exc.	exc.	exc.	exc.	exc.	exc.	exc.
v) recurrence	2	0	0	7	1	1	no
vi) age	38	69	66	48	59	43	58
vii) sex	M	F	F	M	F	M	M
viii) follow up period	$6\frac{1}{2}$ years	4 months	5 years	20 years	9 months	20 years	7 months

Although infiltrating angiolipoma and lipoma share many similarities, these authors believe that enough differences to classify these two lesions into two distinct clinico-pathologic entities exist. With this in mind, they looked at several aspects of the clinical presentation, that is, age of onset, recurrence rate, sex, symptom duration and site of occurrence, and analyzed what they considered "acceptable cases" from the literature with their own cases. A total of 27 tumors

was analyzed. From the 13 cases of infiltrating lipomata, they noted the following:

TABLE V Dionne and Seemayer: Results of Clinical Studies

patients' mean age at diagnosis	50.0 years
sex ratio	1:1
symptom duration prior to excision	6 years
site predilection	lower extremities
recurrence rate	62.5% with follow-up period of 4 months to 20 years.

A total of 14 infiltrating angiolipomata were likewise analyzed with the following results:

patients' mean age at diagnosis	35.5 years
sex ratio	1:1
symptom duration prior to excision	18 years
site predilection	none
recurrence rate	50% with follow-up period of 3 months to 13 years.

In this analysis, the results concerning age of onset were obtained by excluding three cases where the lesion was present at birth. Of these three cases, one was an infiltrating angiolipoma, the rest were infiltrating lipomata.

Enneking and Stimpson discussed in their study the difference found between infiltrating and <u>non-infiltrating</u> angiolipoma. Non-infiltrating angiolipoma are found in subcutaneous tissue as multiple, well encapsulated lesions that do not recur after enucleation. Klem and Rasanen pointed out

in separate studies that these encapsulated angiolipomata were painful.[24, 39] They also noted a familial disposition and postulated trauma on rest cells as the etiological explanation. Howard and Helvig in a more current study of 261 cases, concurred with these findings and demonstrated that the trunk and lower extremities were the most frequent sites of occurrence.[19] Because the mean age of onset was 17 years of age they speculated that the hormonal changes of adolescence coupled with trauma on rest cells gave rise to encapsulated angiolipoma in the predisposed individual.

Enneking has commented that it is possible for these lesions to start as encapsulated and subsequently progress to become unencapsulated and infiltrating. This phenomenon is apparently demonstrated in a case report by Bradley and Klein of a 38 year old male who presented with a nodule in left shoulder of 13 years duration. A tender mass was found centered about the spine of the scapula. The overlying skin was normal. Radiographic study of the left scapula showed an irregular cystic multilocular bone defect involving the acromion and body of the scapula. At surgery, an encapsulated fatty tumor with pedunculated appendages was found lying in the infraspinatus fossa. These pedunculations caused erosive changes in the scapular as well as the acromion. The mass was enucleated without recurrence for six years post-operatively. This case was unusual in that it represents the first instance of an "encapsulated" angiolipoma observed to cause bony erosion.[5]

Case Reports

Two case reports will be presented. The first case is a true example of infiltrating angiolipoma. The second case, which is an example of infiltrating intramuscular lipoma is added to underscore the possible difficulty in arriving at the diagnosis of angiolipoma.

Case 1 J.G. is a 14 year old male who has had multiple admissions to Yale-New Haven Hospital for recurrent swelling in the left knee.

This patient's history dates from the age of 3½ years when his parents noted swelling of his left knee. They also noticed that he walked stiffly with an abnormal gait. The joint was never painful. An aspiration of the joint was performed, but no fluid was obtained. X-Rays were interpreted as showing a possible tumor and on 12-3-65 the patient was admitted to the hospital.

Past medical history revealed that the patient was a product of an 8 month gestation period. The labor was induced prematurely because the mother was toxemic. He was discharged after three days in good health.

At age 5 weeks, he was admitted to another hospital because of vomiting. He was found to have pyloric stenosis for which he underwent pyloroplasty. At age 11 months, he was admitted because of an enlarged head circumference (>97 percentile), hypotonia and generalized motor retardation.

A presumptive diagnosis of hydrocephalus was made. Because the ventriculograms were normal, this diagnosis was changed to arrested hydrocephalus. The patient has never developed any neurological or intellectual deficits.

Family History: included no history of relevant diseases.

Review of Systems: except, as indicated elsewhere, was within normal limits.

On physical examination, the knee was slightly enlarged with some degree of floculation and it was warm to touch. The range of motion of the knee was normal.

Determination of the Alkaline Phosphatase as well as the serum calcium and phosphate was done to rule out bony involvement. They were found to be within normal range. Juvenile rheumatoid arthritis was also included in the differential diagnosis. The patient, however, was seronegative for rheumatoid factor.

At surgery a mass of fibro-adipose tissue was excised from the fat pad of the left knee. The essence of the pathologic findings was as follows: A tissue specimin, in formalin, of predominantly yellow adipose tissue that weighed 22 grams was received for investigation. Microscopic examination revealed fibroadipose tissue with many small blood vessels within the tissue substance. Although most vessels were small, they varied in size. The histologic diagnosis was compatible

with angiolipoma with focal scarring. (Figures 1-3) The patient had a satisfactory post-operative course and was discharged after the tenth hospital day.

He was being followed in the orthopaedic clinic when the knee again became edematous and warm. He was admitted on 11-6-66 to rule out juvenile rheumatoid arthritis.

On physical examination, the patient was afebrile. He had no signs of systemic juvenile rheumatoid arthritis, that is, rheumatoid rash, lymphadenopathy or hypatosplenomegaly.[45] The left knee, however, had a medial enlargement that was soft and warm. There was also a 2 cm. of atrophy of the left quadriceps.

At surgery, a synovectomy was performed. Hypertrophic synovial tissue was removed from the area of the lateral condyle of the femur. The tissue, in formalin, that was received by the pathologists showed a composite of multiple white fragments totalling less than 5 c.c. The histologic examination showed hyperplasia of the synovial tissue. The sub-synovial tissue contained sparse chronic inflammatory infiltrate and many dilated vessels. The diagnosis was chronic synovitis.

The patient had a benign post-operative course and was discharged after the twelfth hospital day.

While at home, the patient began to walk with a limp. His left knee was noted to be more swollen; however, it was without pain. The patient was admitted on 11-23-67 for ex-

cision of a probable recurrent tumor.

After his discharge in 1966, he was followed until the time of the current admission in the orthopaedic clinic. A leg length discrepancy of 2.7 cm. was noted which was believed to be secondary to the hypervascularity seen about the distal femoral epiphysis during the synovectomy of 11-7-66. This increased circulation and warmth was thought to be responsible for the rapid epiphyseal growth of the left knee.[23,25,30,50,51] The patient was given a right shoe lift to correct the length disparity.

A flexion contracture of the left knee developed and the existing atrophy of the left quadriceps progressed. He was treated with physical therapy to strengthen his left quadriceps. For the flexion contraction, he was fitted for a posterior knee splint to be worn at night.

Because the left knee remained enlarged and warm with respect to the right knee, the patient was thought to have rheumatoid disease and was referred on 7-7-67 to the Pediatric Collagen Clinic.

The patient was again found to be seronegative for rheumatoid factor. Radiographs of both knees revealed an effusion in the left knee with suprapatellar extension. A size discrepancy between the bones of both knees was also noted. That is, the left was larger than the right. However, there

was no radiographic evidence to suggest rheumatoid arthritis; that is, no osseous destruction or joint narrowing.[42] (Figure 4) Therefore, it was thought the patient had angiolipoma and not juvenile rheumatoid arthritis.

On physical examination, the knee was red and swollen. A slightly tender, soft tissue swelling was present in the medial aspect of the knee. However, there was no evidence of fluid, pulsations or bruits.

During this admission, the patient was noted to have microscopic hematuria. The results of the investigations carried out are the following:

Urinalysis: 1-10 erythrocytes/high power field

Addis Count: RBC = 75,000,000

WBC = 5,000,000

Casts (granular) = 50,000

IVP: filling defect of the right calyceal system

Cystoscopy: within normal limits

ASLO < 50 todd units

L.E. Test; ANF; Genetic Screening = Negative

It was decided that if the hematuria continued the patient would be readmitted for renal biopsy.

At operation, a mass was dissected from the suprapatellar pouch. Both menisci were resected because of infiltration by the lesion. The specimin received by the pathologist for ex-

amination was a portion of menisci with attached dense fibroadipose tissue measuring 7x4x3 cm.; and multiple segments of tan yellow to pink tissue averaging 4x2x1 cm. The microscopic examination revealed portions of striated muscles; portions of collagenized connective tissue suggestive of tendon and portions of synovial tissue. The synovial tissue contained focal collections of hyperplastic synovial epithelium. There were regions noted about the synovia consisting of fibroadipose tissue with large, prominent vessels similar to those seen in previous biopsies. The diagnosis was angiolipoma.

The patient had no complications post-operatively and was discharged after the tenth hospital day.

Because of persistent hematuria, he was admitted for renal arteriogram and renal biopsy. His blood pressure was 90/60. He had no sign of edema. The B.U.N. and the Creatinine Clearance were 13 and 70 respectively. The urine culture was negative. Renal arteriogram demonstrated no malformation of either kidney. Microscopic examination of the renal tissue revealed acute glomerulitis. During the arteriography, dye was also injected into the left iliac region and films were taken of the left knee. However, no abnormal vascularity was demonstrated.

He tolerated the procedure well and was discharged after the fourth hospital day.

The patient was admitted on 7-8-70 for femoral arteriography because of reappearance of leg swelling.

During the time interval between 1968-1970, the patient had follow-up evaluation in the orthopaedic clinic. The patient had a persistent flexion contracture of the knee which lacked 20° of full extension. It was decided to put the knee in a cast with gradual wedging. At the end of three weeks when this form of therapy was terminated, the knee had gained 15° of knee extension.

In 6-6-69, a determination of bone age of both knees were done. The bone ages of the right and left knees were found to be respectively 8 and 11 years according to the standards of Pyle and Hoerr.[17,37,38,41] The patient's chronologic age was 7 3/12 years.

Six months prior to this admission, the knee began to increase in size as well as to become warmer. He was seen again in the orthopaedic clinic two months prior to the current admission. A palpable mass was found in the left anterior distal thigh. Other positive physical findings included an enlarged left patella.

Arteriography revealed a large vascular mass that was supplied predominantly by the deep femoral artery. Three areas of vascularity having changes consistent with those seen in tumor vessels were noted. The vessels were large, tortuous

and irregular. Pooling of contrast media and early venous filling were other findings suggesting tumor vessels.

Following arteriography, he was discharged on the third hospital day because of exposure to mumps. (Figure 5)

Because arteriography supported the suspicion of recurrence, he was subsequently readmitted on 7-28-70 for elective surgery. A tumor that was grossly angiomatous in nature was exposed. This lesion had three independent foci of involvement: the middle third of the rectus femoris, the patella region and the knee joint. The proximal, medial and lateral aspects of the quadriceps showed no involvement with the tumor. Therefore, they were spared when the radical excision of left quadriceps was performed. Included in the resection was the patella and its tendons. The final specimin weighed 450 grams and measured 23 cm. in length. Above the patella there was a discrete film lipoma mass throughout which were scattered 1-2 mm. diameter dilated vessels. Three fresh specimins of fibroadipose tissue were also submitted with an average volume of 7 cm. This fibroadipose tissue also contained multiple areas of dilated vessels 1-3 mm. in diameter.

Histologic examination revealed dense connective tissue and skeletal muscle being infiltrated by mature fat cells. The fat cells surrounded many thin walled sinosoidal veins as well as thick walled arterioles. Acute inflammatory cells were ob-

served around the small vessel walls. The fat cells showed no evidence of mitosis or lipoblastic activity. Skeletal muscle was noted to be in various stages of atrophy and scarring. (Figures 6-8)

The patient had a satisfactory post-operative recovery with good healing of the wound. He was discharged after the tenth hospital day.

He was admitted on 11-24-70 for repair of a fractured left femur from a fall he sustained during athletic activity at school. Physical examination revealed a warm and swollen knee with a limited range of motion because of pain. An impacted transverse fracture of the distal femoral shaft was seen on radiography with posterior angulation of the distal fragment. A diagnosis of pathologic fracture was entertained pre-operatively; however, no evidence of this was seen when open reduction was done. The patient was discharged on the second hospital day with a walking spica cast.

A month after open reduction was done, the cast was removed. Radiographs showed good healing and alignment at the fracture site.

Following the discharge in 1970, he has been an outpatient at the orthopaedic clinic. On 5-4-71, five months after the removal of the cast, he was found to have a painless ankylosis of the left knee. However, when he returned a year later

in 1972 for evaluation, he had regained some motion of the left knee. He was continued on physical therapy consisting of active resistant exercises for the left leg. On subsequent follow-up evaluations in 1973 and 1974, the patient showed no evidence of recurrence of tumor.

On 1-8-76, he presented to the orthopaedic clinic for a follow-up evaluation. He offered no complaints of pain or discomfort.

On physical examination, the patient was an asthenic but healthy appearing adolescent male who wore a left posterior knee splint. A long curvilinear healed incision was seen from the anterioinferior iliac spine down to the medial aspect of the knee. There was obvious disparity in the size of both thighs. (Figures 9-10) Full extension of the left knee was possible but flexion lacked 60° of its range of motion. Other positive findings observed was a healed pyloroplasty incision on the abdomen.

Laboratory- Orthoroentgenograms revealed diffuse demineralization of distal left femur and the beginning of bilateral epiphyseal fusion. Leg lengths were obtained and showed a discrepancy of 3.2 cm.; the right leg was 96.1 cm. and the left leg was 99.3 cm.

Impression- There was no clinical evidence of recurrence.

Plan- The plan is to continue observation for recurrence.

Case Summary

The relevant aspects of the history of the first case are as follows: J.G. is a 14 year old white male who had been admitted four times for excision of a recurring mass in the left lower extremity. The patient had a painless swelling that was warm to touch. Subsequently, local gigantism of the left knee was observed both on physical and radiologic evaluations. Other pertinent laboratory findings included a normal serum calcium and phosphate. At age 3½ years, the patient had 22 grams of tumor excised from his leg. Subsequent recurrences required excisions of specimens measuring 5 cc., 92 cc. and 450 grams respectively. The histology was consistent with infiltrating angiolipoma except in the second instance where it showed changes of chronic synovitis. Post-operatively, the patient has done moderately well with some disability of the left knee. He has been receiving physical therapy and wearing a posterior knee splint for stabilization of the left knee. He has been without recurrence of tumor for five years.

The second case does not qualify as a true case of angiolipoma because the characteristic vascularity was not found on histologic examination. However, the second case is presented because it was initially diagnosed as an infiltrating angiolipoma. Subsequent histologic review proved this to be

incorrect. This case represents an example of the problematic nature of differentiating these two tumors.

Case 2 M.M., a 13 year old white male, has had three resections for a recurrent mass in the back.

The onset of this patient's disease was at the age of six months when he presented at another hospital for excision of a soft tissue mass in the area of the right scapula. The histological diagnosis at that time was a lipoma.

He was free of any recurrence until the age of 8 years when he discovered a small mass in the region of the previous surgery. The patient denied any pain. Because the tumor had increased in size over the following 3 years, the patient was admitted on 8-18-74 for surgery.

The past medical history revealed that the patient was a product of a full-term gestation. His growth and development was within normal limits. The family history was non-contributing for history of relevant diseases.

The physical examination was normal except for the presence of a large subcutaneous mass at the lower edge of the right scapula. There was extension of the tumor towards the axilla and up the vertebral border of the scapula. The mass was soft and non-tender.

Chest films showed elevation of the right fifth rib with spreading of the fourth and sixth ribs. There was also

rotation of the fifth costotransverse junction and a 15-20 degree scoliosis centering at the fifth thoracic vertebra. A pre-operative diagnosis of scoliosis secondary to neurofibromatosis was entertained.[6,20,26] (Figure 11)

At surgery, a large lipomatous growth that was primarily subfacial in location was discovered. This lesion of approximately 1½ inches in thickness extended across the mid-line at the spinous process of the eighth thoracic vertebra. This lesion also invaded other deep structures, namely: the paraspinous muscles of the third through the sixth thoracic vertebrae, and the intercostal muscles and finally the periosteum of the fourth and fifth right thoracic ribs. A sub-total excision was performed with most of the paraspinous muscles being spared.

The main tumor mass measured 23 cm. x 7 cm. x 2 cm. and contained on its surface periosteum of the patient's rib. The lesion on histologic examination was reportedly composed of trabecular masses of mature adipose cells and aggregates of capillaries, arterioles and veins of various caliber. This fatty tissue interdigitated with soft tissue and skeletal muscle. A histologic diagnosis of infiltrating angiolipoma was made. However, upon review of the slides, this diagnosis was changed to infiltrating lipoma. The review confirmed the presence of adult fat tissue infiltrating muscle but it differed from the original report with respect to the vascularity within the le-

sion. That is, the vessels were found to be normal in number and appearance. The lack of excessive vascularity and neoplastic appearing vessels, therefore, argued against the original diagnosis of angiolipoma. (Figures 12-13)

The patient had no difficulty post-operatively and was discharged after the fourth hospital day.

On 3-5-75 the patient was readmitted because of tumor recurrence at the site of the incision.

The patient noted one month prior to admission rapid enlargement of the mass. He denied pain in the area of the mass.

On physical examination, a large soft mass elevating the scapula was found. It extended across the midline posteriorly and extended to the axilla anteriorly. There was no branchioplexus involvement noted.

Radiographic studies revealed no progression of the patient's existing scoliosis. Abnormalities of the fifth thoracic vertebra and its rib were described. The rib was enlarged with respect to the other ribs and the vertebral body was slightly higher on the right than on the left. Radiographic views of the right scapula showed soft tissue swelling without evidence of bony destruction. (Figure 14)

At surgery, the lesion was removed from under the scapula along the latissimus dorsi and teres minor. The entire serratus anterior was resected. This resulted in a winged scapula.

The entire paraspinal musculature had diffuse involvement with the lesion which was not resected.

A fresh specimin measuring 15 x 13 x 2 cm. composed of fibroadipose and muscle tissue was sent to Pathology. It was at this time that microscopic review of previous slides was undertaken. Histologic examination of the current slides revealed similar findings, that is, infiltration of skeletal muscle was noted to be in the various stages of scar formation and degeneration. There were large areas of scar tissue which contained normal appearing blood vessels. (Figure 15) The tumor was diagnosed as an infiltrating lipoma of the intramuscular type. He had a good post-operative recovery and was discharged on the sixth hospital day.

Following discharge, he was referred to the Radiotherapy department on 3-12-75 for evaluation. This form of treatment was not employed for a variety of reasons. It was felt that the risks of worsening the patient's scoliosis by destroying the growth centers or inducing neoplasm were not justified in this patient since the tumor was benign.[2,9,21,22,28,35,43] It was decided to manage any recurrences surgically.

On 2-13-76, the patient returned for follow-up evaluation in the orthopaedic clinic. The patient's parents noted a small swelling of two weeks duration at the area of incision. The patient denied pain.

On physical examination, the patient was a thin adolescent male. A long right-sided thoractomy scar was apparent. At the lower junction of the thoractomy scar was a palpable swelling. There was good shoulder function with less winging of the scapula. The rest of the physical examination was unremarkable.

Laboratory- Radiography revealed no evidence of progression of the scoliosis.

Impression- The swelling was suggestive of recurrence; however, this may represent loculation or a hypertrophic scar.

Plan- The plan was to follow the patient at 6 month intervals for 5 years to note any recurrence of tumor formation.

Case Summary

The main features of the second case are as follows: M.M. is a 13 year old white male who has been admitted three times for a recurring mass in the right upper back. On physical examination, he had a painless soft tissue mass that extended across the midline of the back posteriorly and extended to the axilla anteriorly. Radiographic findings when the patient was 11 years revealed mild scoliosis and an enlarged right fifth thoracic rib. Subsequent recurrences required excision of masses measuring 27 x 10 x 5 cm. and 15 x 13 x 2 cm. respectively. An initial histologic diagnosis of infiltrating

angiolipoma was incorrectly made. However, the final interpretation was that the specimin had features consistent with intramuscular infiltrating lipoma. Post-operatively, the patient has done well with only winging of the right scapula. He has been without recurrence of tumor for one year.

Discussion

A. Clinical Presentation

The most remarkable presenting symptom of infiltrating angiolipoma is pain.[10] However, this has not been borne out by the case presented here. Likewise, the symptomatology of pain does not appear to be found universally in the cases studied. With the inclusion of our case, 14 cases of infiltrating angiolipoma have been reported. Davis' case, which was incorrectly interpreted as infiltrating angiolipoma in the 1971 study of Dionne, is excluded because it does not fulfill the histologic criteria. There is no mention of vascularity in Davis' histologic description of the lesion. From the 14 above cases, 10 (71.5%) have presented with this symptom.

It may be postulated that pain may be caused by the spread of the tumor into muscle secondarily causing pressure on sensory nerves. However, other mechanisms of pain may be contemplated. Pain may, for instance, be secondary to congestion within the vasculature of the tumor. (Figures 3 and 6) This congestion may stem either from external trauma, from growth of tumor within resistant tissue or from vascular obstruction caused by phleboliths. Enneking and his associates observed that the pain as a rule is not present initially at the onset of tumor growth. Thus, it is possible that the

development of vascular congestion over time might be an explanation for the delayed onset of pain in these lesions.

The etiology of the disease is not clear. Some investigators have used the mean age of onset to postulate the etiological nature of infiltrating angiolipoma. As previously mentioned in the review of the literature, Enneking and Stimpson obtained from their respective series contradicting results with respect to the mean age of onset. Enneking et al. found this lesion to be concentrated in the first three decades and hence postulated that the tumors were hamartomas or congenital malformations. Although Stimpson was able to demonstrate a later age of onset from his series which were concentrated in the sixth decade, he did not offer any hypothesis about the etiology. Since it is possible for this lesion to occur at any age, a more attractive hypothesis, we believe, is that sequestered multivalent mesenchymal tissue may indeed be responsible for the genesis of these lesions.

There is no controversy regarding the skeletal muscle being the prime target organ of infiltrating angiolipoma. However, secondary involvement of adjacent bony structure is a positive finding in 2 of the reported cases, as well as in J.G., the case reported here. Bone invasion by infiltrating angiolipoma produces three important consequences. Firstly, the irritative effect of close contact of the lesion with

bone will eventually manifest itself radiologically by periosteal proliferation and thus raise suspicion of bone malignancy. One of the cases reported by Enneking and his associates did indeed demonstrate this radiologic finding (Table II, Case 3). Secondly, the erosive invasion of these lesions can bring the eventual atrophy and death of bony tissue. Again, Enneking et al. offers a case that illustrates this destructive feature of lipomatous growth (Table II, Case 6). Thirdly, because of increased vascularity inherent in angiolipoma, hypertrophy of the affected part ultimately results. The leg length discrepancy of the case of this study demonstrates nicely this phenomenon of localized gigantism.

The clinical feature of local gigantism is an important finding because its etiology may be related to the possible hypervascularity of the tumor on the region. Khaw and Dalik have proposed a classification of localized gigantism of the extremities. The classification is based upon the predominant hamartomatous malformation in the affected area. The lesion may be either of soft tissue or of vascular tissue, with the latter being characterized by hemangiomatosis and A-V fistulae (Kippel-Trenaunay Syndrome).[23] Local or partial skeletal hypertrophy may be part of a systemic disease, a distinct clinicopathological entity characterized by nevi, precocious varicose veins and osseous hypertrophy of the affected part or

a form fruste of any one of these findings. The osseous hypertrophy could be due to the tortuous vessels through which there would be an increased blood flow.[25] Other cases have been reported in which hemi-hypertrophy was found associated with tumor-like vascular overgrowth as well as concomitant nevi.[49,50] Anatomically, the association of specific regions of A-V shunting and arterial tortuosity have been demonstrated by angiography[30] and necroscopy.[25] Despite the fact that hypervascularity appears to be associated with local gigantism, it is interesting to note that only the case of infiltrating angiolipoma presented in this paper had local gigantism. None of the literature cases of infiltrating angiolipoma report this finding.

The location of involvement as a factor in the clinical presentation of infiltrating angiolipoma is an issue of debate between workers like Enneking and Dionne. The former has found the lesion predominantly in the lower extremities. Our personal case with infiltrating angiolipoma is compatible with this clinical presentation. The results obtained by Dionne who concluded that there is no site predilection support the idea that the lesion can theoretically occur wherever voluntary muscle is found. It is probable that Dionne's conclusion is valid since it is based on a larger series composed of 6 cases reported by Enneking and 7 cases reported by Stimpson.

The incidence of this lesion is equally shared by both sexes. Of the 14 cases of infiltrating angiolipomata studied, 7 were males. However, with so few cases in the literature, the basic clinical characteristics of this tumor have not yet been thoroughly delineated. Hopefully, more experience with it will be documented so that more definitive answers can be found.

B. Differential Diagnosis

The differential diagnosis of infiltrating angiolipoma has been problematic because of its rarity and the depth of location of the tumor. The relatively more frequently encountered lesions from which infiltrating angiolipoma should be distinguished are listed as follows:

i) hemangioma
ii) hematoma
iii) liposarcoma
iv) tuberculous myositis
v) fibrous myositis
vi) hydratid cyst
vii) neurofibromatosis
viii) arthritis

Shallow et al. recommend aspiration or exploratory puncture be done as an initial diagnostic procedure in differentiating

the above entities.[46] If the lesion is a primary hemangioma, aspiration of the lesion will yield bright red systemic blood. In addition, the hemangioma will characteristically resume its original size at the termination of aspiration. If dark old blood is obtained on aspiration then it is more than likely indicative of hematoma. Aspiration of blood is possible in some vascular sarcoma, however, the patient may have some signs of systemic malignancy if metastasis has occurred. Patients with infiltrating angiolipoma show no signs of systemic malignancy.

Muscle swelling of infectious etiology are encountered in tuberculosis and in hydratid cyst. Exploratory puncture of a tuberculous myositis will yield casseous material that is positive for acid-fast bacteria. The diagnosis of hydratid cyst is made with a positive Casoni intradermal test and an aspirate of yellow fluid with the characteristic hooklets.

It is possible for a presumptive diagnosis of neurofibromatosis to be made if there is coexisting scoliosis secondary to the erosive effects of adjacent lesion. Neurofibromata tend to be firmer in consistency and sharply demarcated from the surrounding tissue. Other factors present such as positive family history, cafe au lait spots and axillary freckles will strengthen the clinical impression of neurofibromatosis.[26]

Finally if the lesion is in the vicinity of a joint it is likely that an arthritic process will be suspected, as in our case reported below here. Helpful diagnostic work-up for the arthritides are arthrocentesis, blood studies, radiography and biopsy.[42]

C. Pathology

Stimpson observed from his study that the gross morphologic appearance of these lesions were, in the majority of cases, similar to simple lipomata. Therefore, there may be no conspicuous sign of underlying angiomatous pattern such as tortuous vessels on the tumor surface or excessive hemorrhage during extirpation of the mass. Of the 14 cases of angiolipoma studied, only 5 (28%) cases had obvious morphologic findings of abundant and abnormal angiomatous pattern. It is thus correct to conclude that the diagnosis of infiltrating angiolipoma is a histological one.

Histologically, adipose tissue and blood vessels are the two predominating elements. The adipose tissue is composed of mature fat cells whose growth is not by expansion as in the usual benign tumor but by infiltration. The vessels, however, are morphologically neoplastic in appearance, and present in greater number than those found in the lipomatas. The distribution of these vessels is also atypical; that is,

they are found within the fatty tissue itself either as aggregates of capillary, artery and vein or as individual vessels surrounded by larger accumulation of fat cells. The vessels themselves are a bizarre collection of thin walled cavernous veins and thick walled arterioles. In the 6 cases where obvious abnormal vessels were present on the tumor surface, microscopic examination revealed a predominance of large cavernous veins. This correlation of gross and histologic morphology may account for the low incidence of infiltrating angiolipomata with striking vascularity on its surface.

Of the two histologic criteria outlined above for infiltrating angiolipoma, only one is shared with the other clinical entity of infiltrating lipoma. That is, they both have in common the presence of adult fat cells of which the growth is by infiltration. Separation of the two entities is based on the criteria of vessel histology. In infiltrating lipoma the vessels are normal and not neoplastic in appearance. There are instances, however, when the appearance of the vessels is changed because of inflammatory reaction. Furthermore, the number of vessels is not excessive and its distribution is primarily within scar tissue of degenerating muscle fiber. This is clearly not the pattern observed in infiltrating angiolipoma where the vessels are ubiquitous in their distribution, be it in fatty tissue or scarring tissue. Thus, we conclude that some of the difficulty regarding the diagnosis

of infiltrating angiolipoma versus infiltrating lipoma stems from correct morphologic interpretation of the vessels present.

D. Management

In cases where extensive growth is not present, most workers believe that the most optimal form of therapy can be achieved by wide local resection. Radical surgery in cases with extensive involvement becomes complicated with mutilation and loss of function of the affected part. However, should contiguous structures like nerves or systemic blood vessels also be involved, most authors would opt for a sub-total excision to be followed by a course of radiotherapy, thereby eradicating microscopic infiltration.

Angiography is useful both in the diagnosis and the management of infiltrating angiolipoma. In this lesion, serial arteriograms will show irregular formation of new vessels, increased rapidity of flow from the arterial to the venous system.[8,44,46] However, before radical surgery is done, most authors agree that angiography should be done to outline the tumor border and to demonstrate communication with systemic vasculature.

Finally, some authors suggest that arteriography may be of value in the assessment of response of vascular tumors to radiation therapy. That is, the progressive disappearance of abnormal vessels noted on the initial arteriogram would provide

an index of response. However, while this form of management is employed in other vascular lesions like hemangioma, this has not yet been tried with infiltrating angiolipoma.[46]

E. Prognosis

The prognosis of infiltrating angiolipoma is characterized invariably by recurrence. Of the cases reviewed, 50% had a complete recovery, that is, no recurrence occurred after the initial extirpation during a follow-up period of 12 months to 4 years. However, among those who had recurrences, mean recurrence rate of 1.86 was found during a follow-up period of 3 months to 13 years.

The prognosis of the lesion depends on the extent of involvement encountered at the initial excision and the adequacy of this excision. At best, with early diagnosis and extirpation, the patient will be free of recurrence. At worst, multiple resections to control recurrence will eventually lead to functional impairment and deformity of the affected part.

Conclusion

This paper is an analytic review of 14 cases of infiltrating angiolipoma. Thirteen of these cases have been selected from the literature and a new case has been added.

Pain was a variable finding among the cases studied; it was the cardinal symptom in 71.5% of cases. The symptom of pain was more prevalent among patients under the age of thirty. The 64% of cases presenting initially during that time span gave support to the prevailing theory that these lesions are congenital malformations. The lower extremities were found to be involved in 57% of the cases. This is a much lower percentage than the 85% found in the series of Enneking and his associates in 1966. However, more cases have been reviewed since then with more diverse sites of involvement. Therefore, while the lesion has been observed in other locations, the lower extremities appear to be the most common site of involvement.

The skeletal muscle is the prime target organ, but in 3 out of 14 cases (21%), bony involvement and its consequences were noted. In this respect, our case presented with unique findings in the skeletal system that resulted from the lesion. First, there was local advanced skeletal maturation of the knee involved by the tumor. There was also overgrowth of the limb on the side of this knee. Finally, there was localized gigan-

tism of the bony components of the affected knee.

Other characteristic features of the clinical presentation of this disease are a non-sexual predilection, and a negative family history. The differential diagnosis is difficult because of the small number of cases reported. This points to the value and necessity for future detailed reports and review since the basic clinical characteristics of these tumors have not yet been thoroughly delineated.

Because of the lesion's depth of location, it can be confused with other more frequently occurring soft tissue masses. Therefore, a biopsy is needed to make the diagnosis. The gross appearance may vary from that of an hemorrhagic angioma of the skeletal muscle to that of the lipoma. However, the gross morphologic appearance in 82% of the tumors was no different from that of the common lipoma. The definitive diagnosis of infiltrating angiolipoma is made on histologic examination.

Adult fat cells characteristically infiltrate and displace muscle (Figre 8). In addition, the morphology of the angiomatous component is abnormal. The vascularity of infiltrating angiolipoma is distinct in 3 ways. The vessels of the angiolipoma are either thin walled cavernous veins or thick walled arterioles as opposed to the normal sized vessels seen in the lipoma. (Figures 3 and 13). Unlike the infiltrating lipoma, its total number of vessels exceeds the expected

amount necessary for tissue nutrient requirements (Figures 1 and 15). By using the lipoma for comparison, the ubiquitous nature of the vascular distribution of infiltrating angiolipoma can be appreciated (Figures 1, 3 and 17). Since this distinction has not been mentioned in other studies, it will be of diagnostic interest to note whether or not future reports will confirm this finding.

At this time, the most widely used form of treatment is radical surgical excision. It is interesting to note, however, that of the cases reviewed 4 (29%) were treated with radiation therapy in an attempt to prevent recurrence following sub-total excisions. Radiation is a valuable form of therapy. It is usually reserved for conditions in which treatment with surgery would involve severe mutilation. Finally, angiography is advocated by most authors as a useful adjunct in the management of the patient with extensive involvement in order to demonstrate abnormal vascularity and to detect communication with systemic vasculature.

The prognosis of this lesion is variable. This depends largely on the location and the involvement of the lesion. Its infiltrating tendency and 50% recurrence rate require multiple radical excisions in many cases. Therefore, a "cure" is usually achieved only with the sacrifice of some functional muscle mass and thus some impairment and mutilation of the affected part. The tumor can be described as lo-

cally recurrent; however, the cells do not characteristically undergo malignant degeneration nor do they metastasize.

Summary

A careful analytical review of 13 cases of infiltrating angiolipomata (reported in the English literature) has shown this to be a benign process in spite of its high incidence of recurrence. From the review of the literature, most workers have observed the following diagnostic criteria:

A. Clinical Presentation
1) age of onset before third decade
2) negative family history
3) no sex-predominance
4) predilection for lower limb involvement
5) pain in the affected region

B. Pathological Examination
1) infiltration of skeletal muscle by adult fat cells
2) thin-walled cavernous veins and thick-walled arterioles
3) excessive number of vessels in lesion

The case presented in this study, J.G., was found to conform with the above mentioned criteria with the exception of localized gigantism in the region affected by the lesion.

The differential diagnosis for infiltrating angiolipoma is varied, i.e., hemangioma, hematoma, myotosis, neurofibromatosis and arthritis. The most difficult is that of infiltrating lipoma. These two lesions are similar in their gross morphology and in their biologic behavior.

A case presentation of infiltrating lipoma is included to illustrate the difficulty that can arise in differentiating these two lesions. This study has shown that the vessels of infiltrating angiolipoma have a ubiquitous distribution within the tumor mass: i.e., it is found within the fatty tissue as well as the scar tissue. In infiltrating lipoma, the vascularity is confined to the scar tissue. This distinct histological characteristic of infiltrating angiolipoma allows for another possible diagnostic criterion.

I have attempted to show the difficulties arising in the differential diagnosis of infiltrating angiolipomata versus infiltrating lipomata. Because of the rarity of reported cases in the literature, set criteria have not been thoroughly delineated. Further detailed reports and investigations are of paramount importance before any definite criteria can be made of this entity.

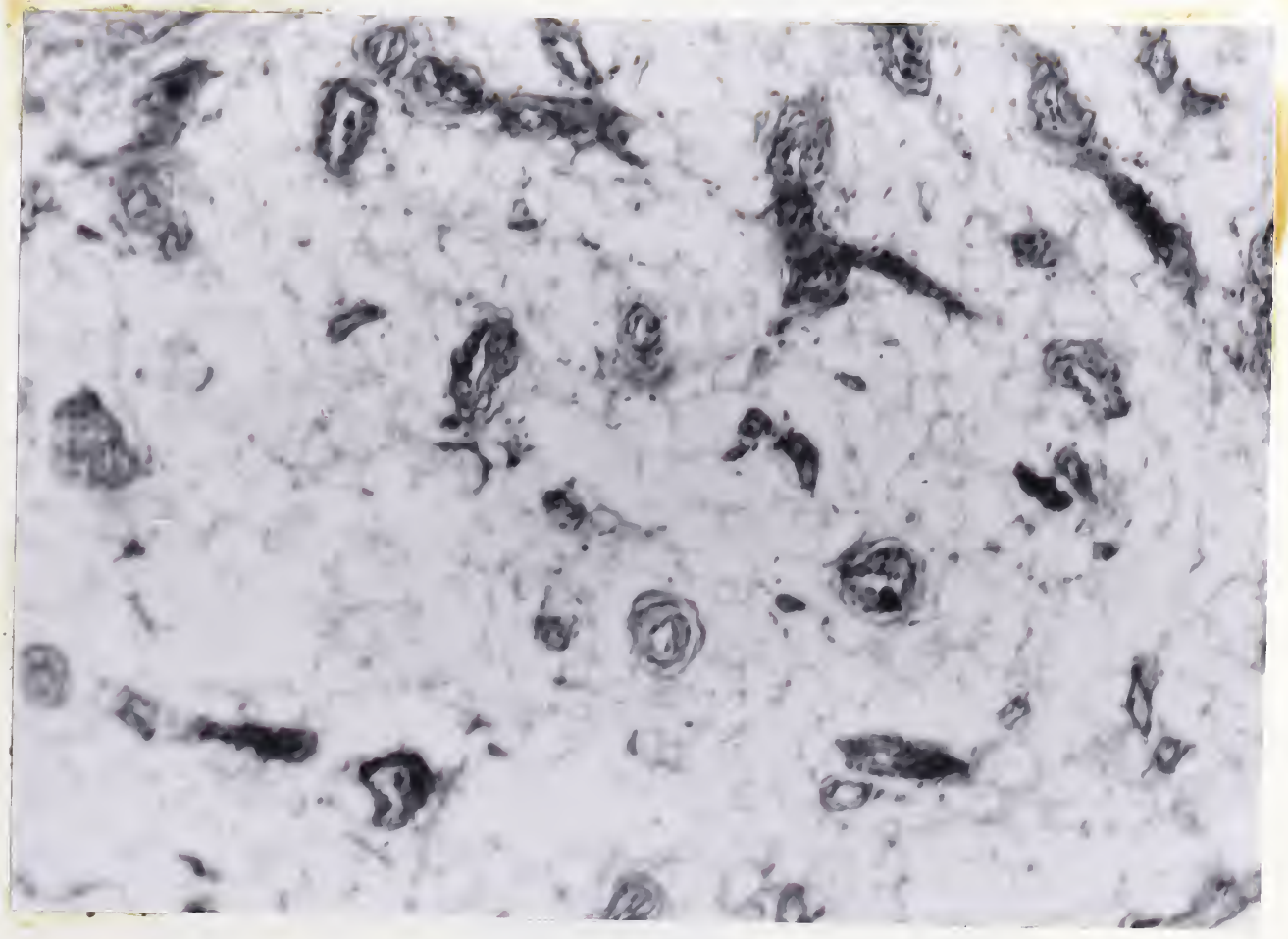

Figure 1, Case 1: Fatty and Vascular Tumor; Small Arterioles, Venules and Mature Fat; X 100; H & E

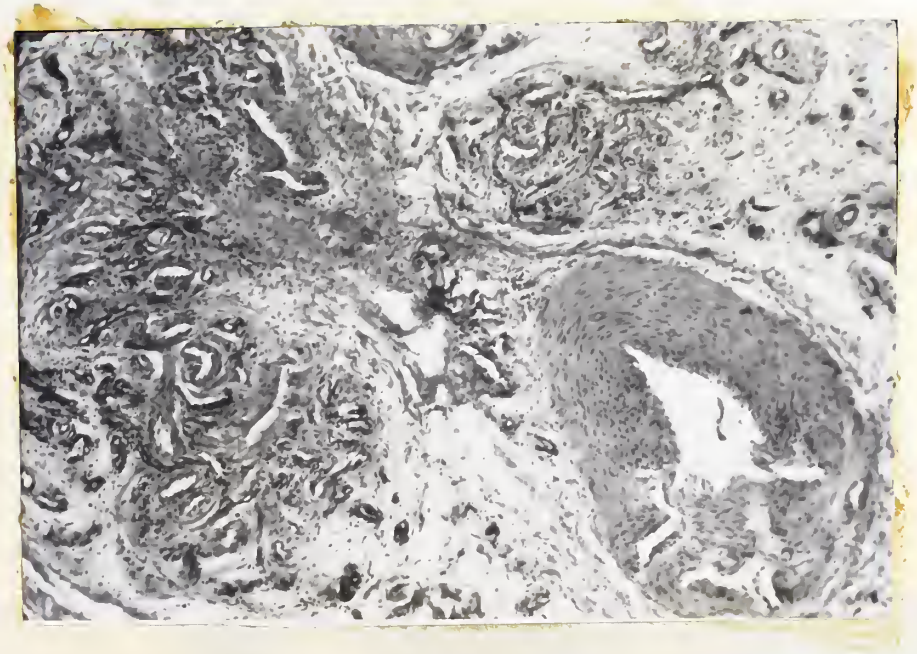

Figure 2, Case 1: Fatty and Vascular Tumor; Large and Small Arterioles, Venules, X 100, H & E.

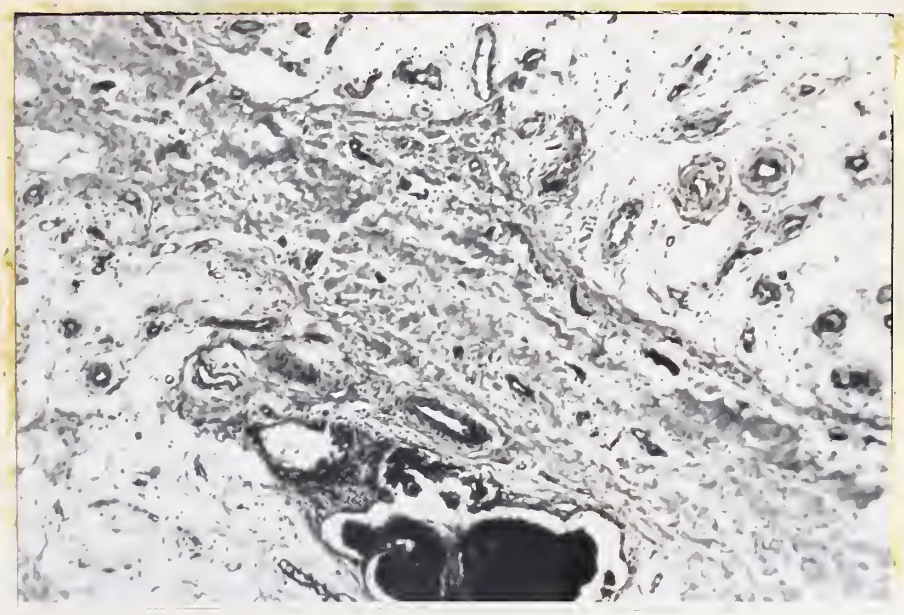

Figure 3, Case 1: Large Congested Dilated Vein and Small Arterioles. Fatty and Connective Tissue Stoma, X 100, H & E.

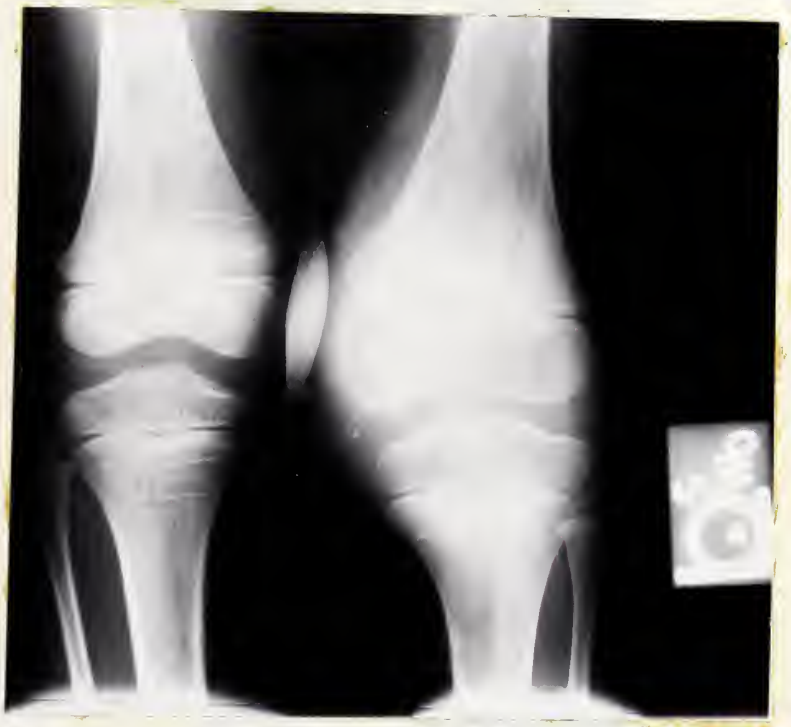

Figure 4, Case 1: Roentgenogram of knees: bones of the left knee are larger than that of the right.

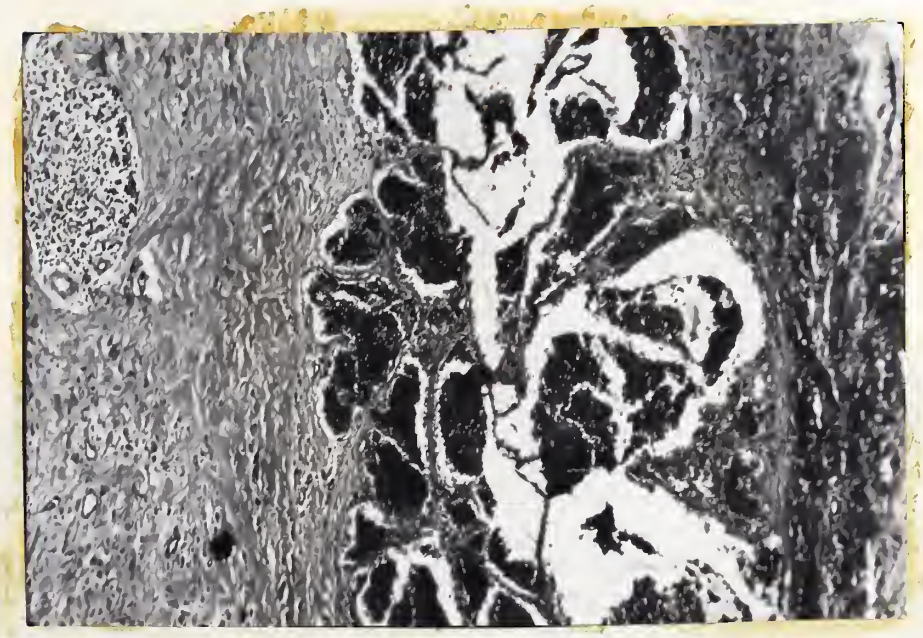

Figure 6, Case 1: Angiomatous portion of tumor; thin walled congested vein indents collagenous stroma, X 40, H & E.

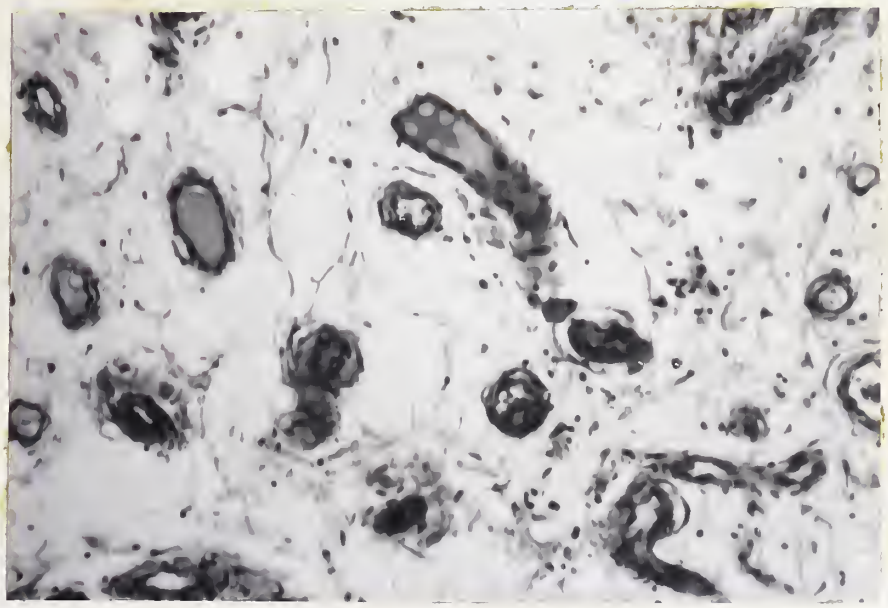

Figure 7, Case 1: Fatty tumor with numerous small blood vessels, X 100, H & E.

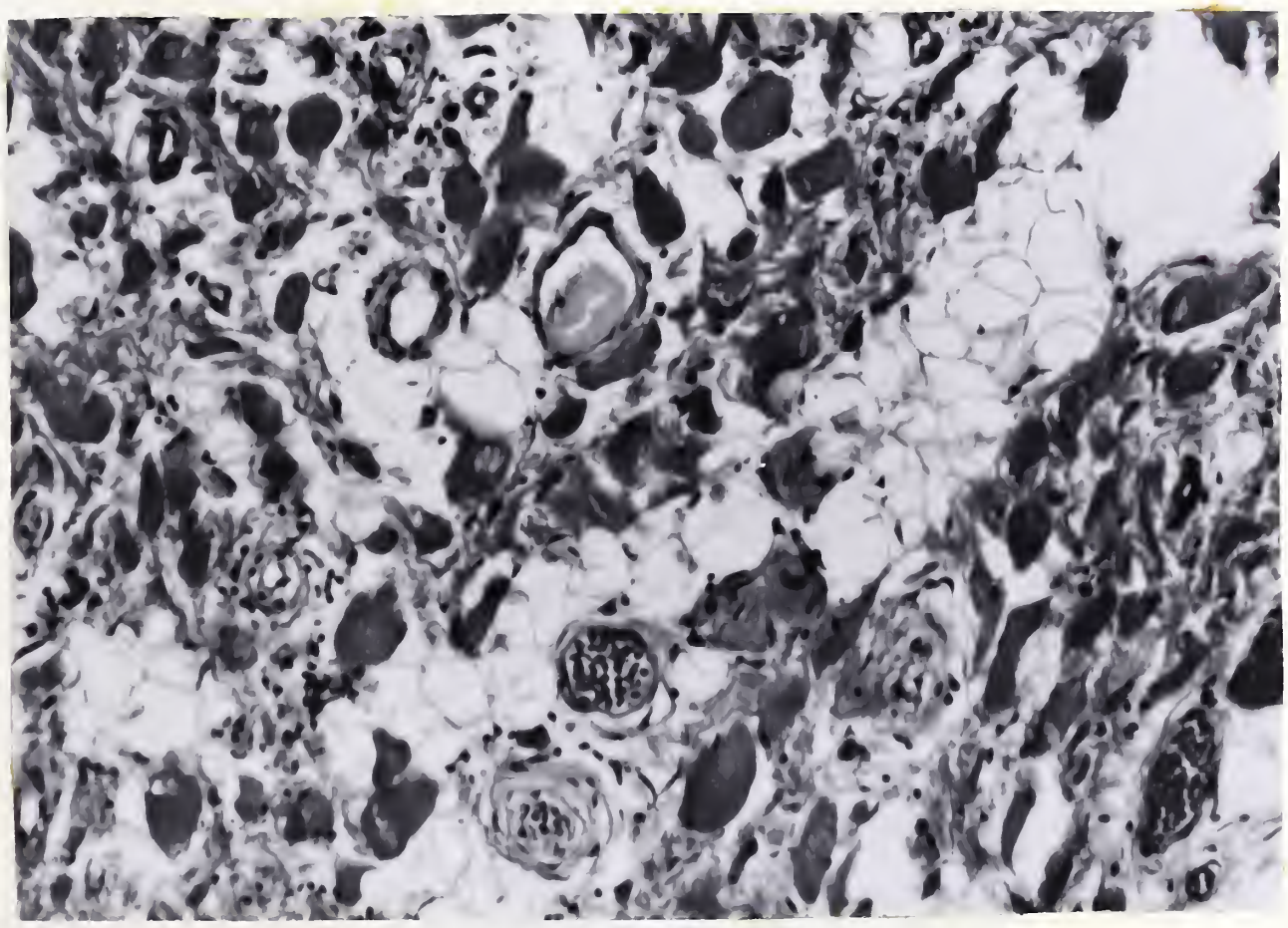

Figure 8, Case 1: Fully developed angiolipoma. Section shows many atrophying skeletal muscle cells, X 100, H & E.

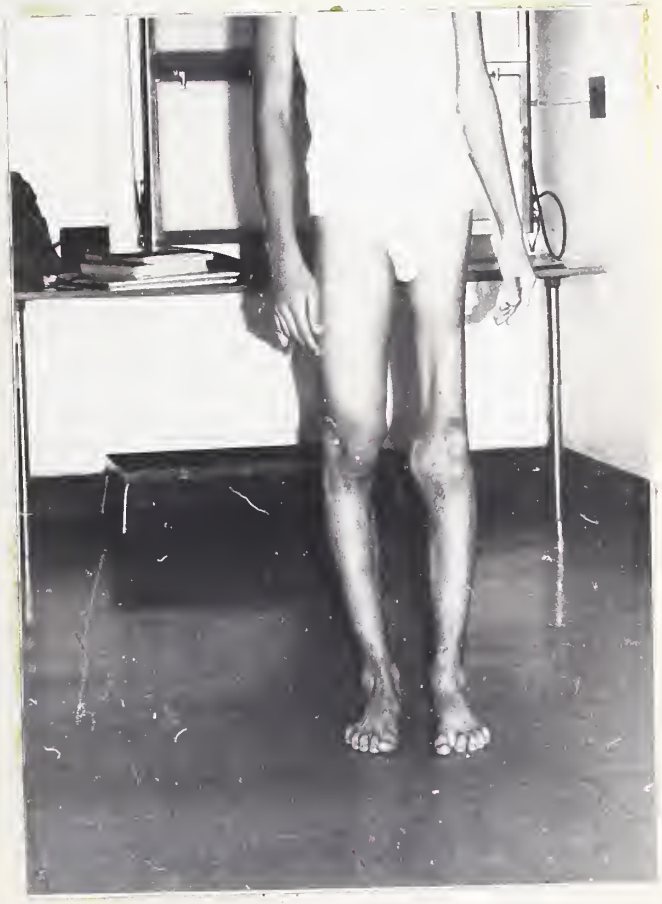

Figure 9, Case 1: Left thigh shows a healed curvilinear scar and decreased muscle mass where quadriceps has been excised.

Figure 10, Case 1: J.G. ambulating with posterior knee splint.

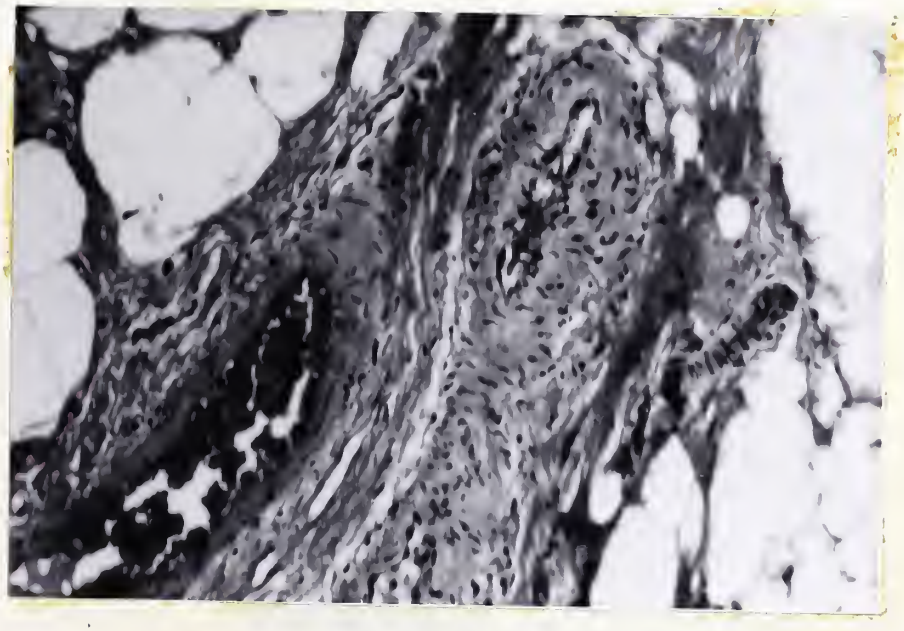

Figure 12, Case 2: Large blood vessels, scar and fatty tissue. Vessels simulate angiolipoma but may be simply reactive. X100, H & E.

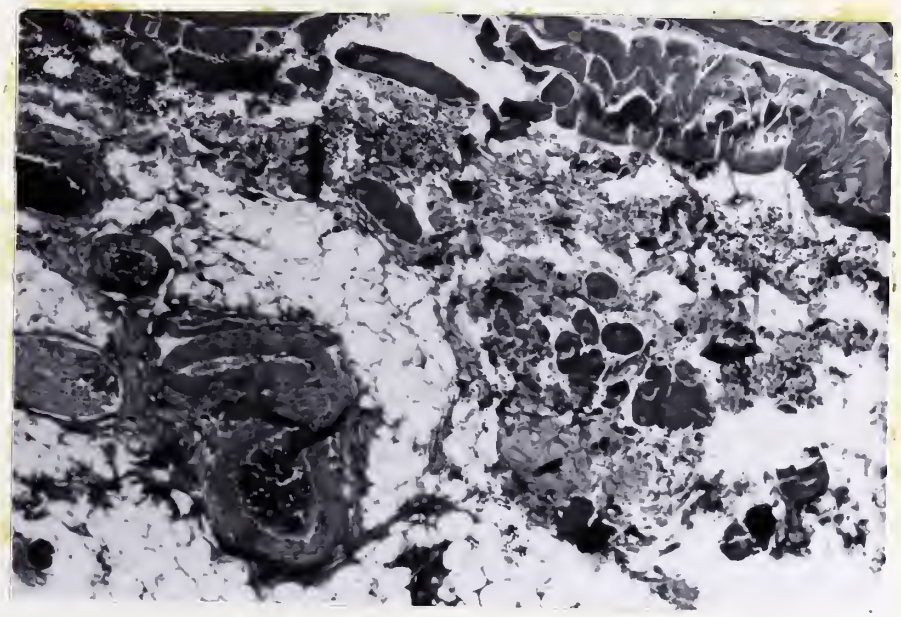

Figure 13, Case 2: Blood vessels, fat, elastic tissue and scar. Muscle and scar being infiltrated by fatty tissue, X 40, H & E.

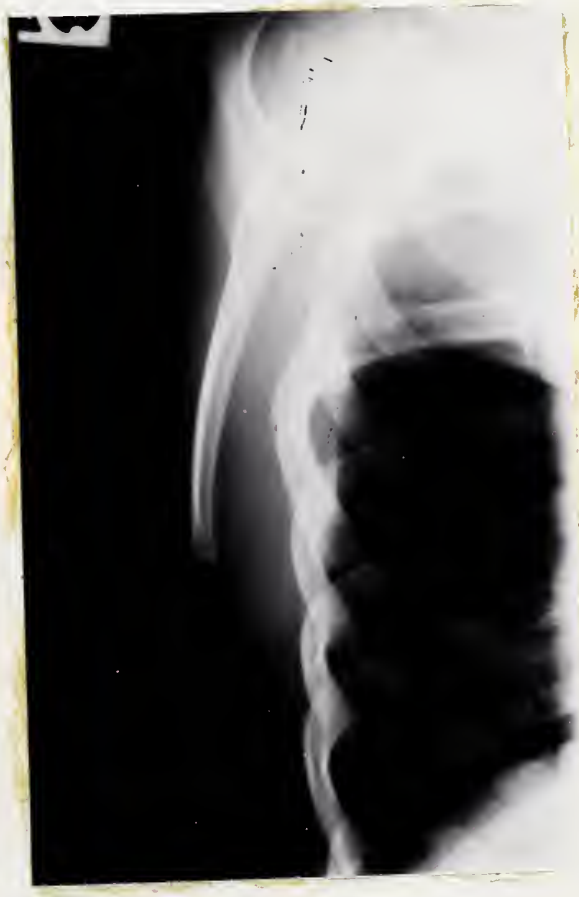

Figure 14, Case 2: Radiographic view of the scapula showing soft tissue swelling.

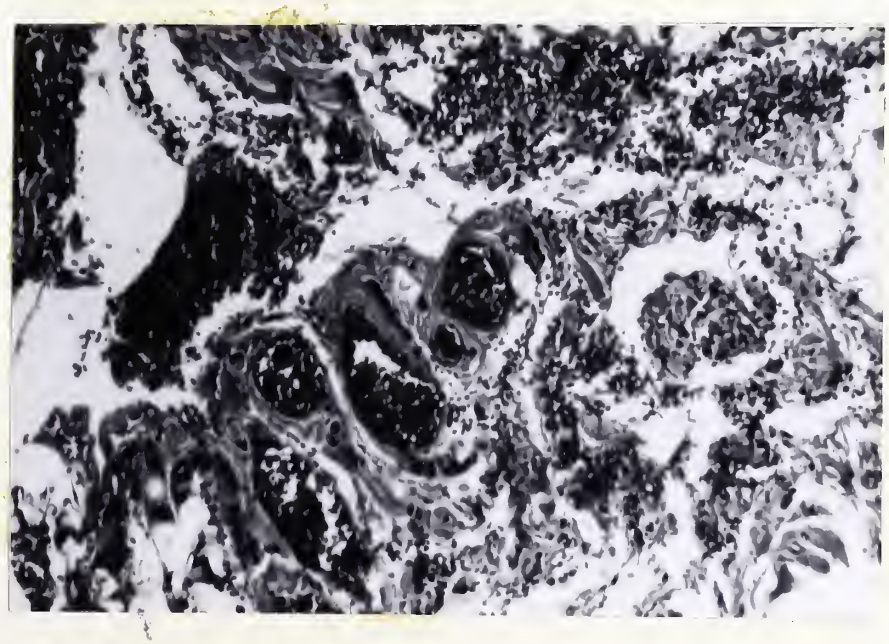

Figure 15, Case 2: Adult fat cells, vessels and scar tissue, X 100, H & E.

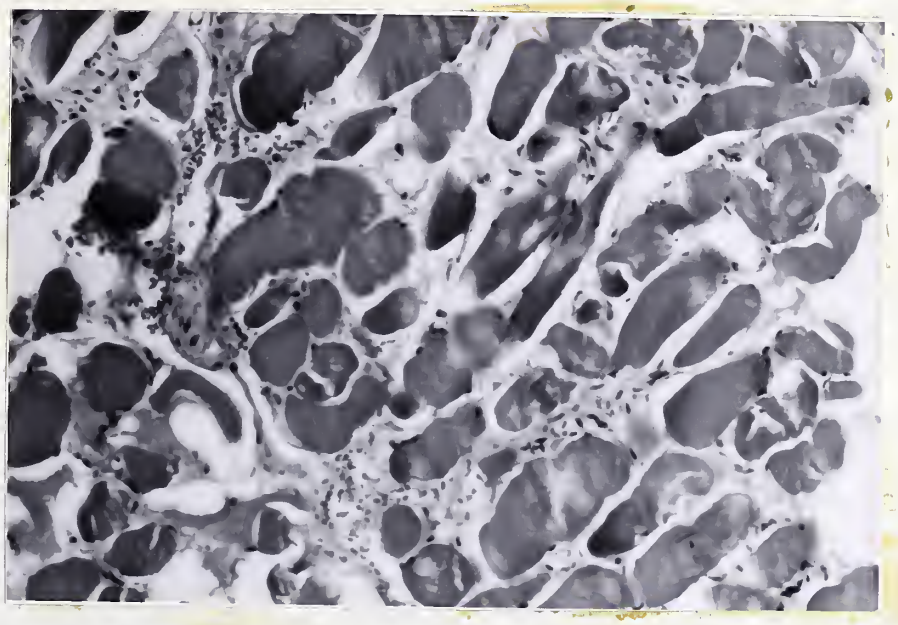

Figure 16, Case 2: Degenerating muscle, interstitial cellular infiltrates, X 100, H & E.

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